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Original article

Predictive factors for sustained remission with stratification by myositis-specific autoantibodies in adult polymyositis/dermatomyositis

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Abstract

Objective. The aim of this study was to clarify predictive factors for sustained remission in adult patients with PM/DM, particularly focusing on stratification by myositis-specific autoantibodies (MSAs).

Methods. A total of 162 adult patients with PM/DM who were followed up for >1 year after diagnosis were retrospectively enrolled. MSAs were evaluated comprehensively in 102 patients whose sera were available. Sustained remission was defined as no evidence of disease activity (active skin rash, active myositis or active interstitial lung disease) for longer than a 6-month continuous period while undergoing myositis therapy or no medication. Clinical data were reviewed in patients' medical charts

Results. The sustained remission rate for all patients was 58% during the median follow-up period at 4 years. With regard to MSAs, the achievement rate of sustained remission among MSA-negative patients was significantly higher than that for patients with anti-aminoacyl-tRNA synthetase ($P = 0.004$), anti-melanoma differentiation-associated gene 5 ($P = 0.037$) or anti-transcriptional intermediary factor 1-γ ($P = 0.013$) antibodies. MSA-negative status (odds ratio 5.84, $P = 0.009$) and absence of severe muscle weakness requiring assistance at diagnosis (odds ratio 43.6, $P < 0.001$) were independent factors associated with sustained remission in multivariate analysis. Cumulative remission rates were significantly higher ($P < 0.001$) in patients with both the MSA-negative status and absence of severe muscle weakness at diagnosis than the others.

Conclusion. MSA-negative status and the absence of severe muscle weakness requiring assistance at diagnosis are independent predictive factors for sustained remission in adult PM/DM patients.

Key words: polymyositis, dermatomyositis, myositis-specific autoantibodies, prediction, treatment outcome, remission

CLINICAL SCIENCE

Rheumatology key messages

- Approximately half of the PM/DM patients achieved sustained remission after induction therapy.
- The absence of severe muscle weakness is strongly associated with sustained remission in PM/DM.
- Myositis-specific autoantibody-negative PM/DM patients have a better clinical outcome than those with other MSAs.

Introduction

PM/DM are idiopathic inflammatory myopathies that affect skeletal muscle, skin, and other organs such as the lungs, heart and joints [1]. The measurement of myositis-specific autoantibodies (MSAs) is highly useful to predict the clinical presentation, treatment response and prognosis in PM/DM [2, 3]. Prognostic factors for patients with PM/DM have been reported as follows: elderly age, male, amyopathic DM, dysphagia, interstitial lung disease (ILD), cardiac involvement and malignancy [3–5]. The MSAs strongly associated with a poor prognosis are anti-melanoma differentiation-associated gene 5 (anti-MDA5) antibody and anti-transcriptional intermediary factor 1-γ (anti-TIF1-γ) antibody, because anti-MDA5 and anti-TIF1-γ are associated with fatal complications developing into rapidly progressive ILD (RP-ILD) and malignancy, respectively [6, 7]. Thus, PM/DM patients with these fatal complications must be managed appropriately to improve their prognosis.

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In addition, control of disease status at low disease activity or remission is also clinically important to improve the activities of daily living for patients with PM/DM. The presence of anti-aminoacyl-transfer ribonucleic acid synthetase (anti-ARS), anti-Mi-2, JDM and less disease damage were associated with good response to rituximab and good outcome for refractory juvenile and adult PM/DM patients according to the Rituximab in Myositis trial [8]. In daily clinical practice, the persistence of Gottron's papules and nail-fold abnormalities early in the disease course were associated with a longer time to remission for patients with JDM [9]. On the other hand, predictive factors associated with sustained remission remain unknown in detail for adult patients with PM/DM in daily clinical practice.

Therefore, in the present study, we investigated the predictive factors for sustained remission in adult PM/DM in daily practice using a single cohort database, particularly focusing on MSAs.

Methods

Patients

This study included 162 adult patients ($\geqslant$16 years old) diagnosed with PM/DM and clinically amyopathic dermatomyositis (CADM) who visited the Saitama Medical Centre, Jichi Medical University between January 2001 and January 2017. The diagnoses of PM/DM and CADM were based on the criteria of Bohan and Peter [10], and the definitions of Sontheimer [11], respectively. Patients with JDM and overlap syndrome were excluded from this study. The follow-up period was defined as the period from diagnosis of PM/DM to either the latest hospital visit or the date of death. The enrolled patients were followed for $>$1 year after induction therapy for PM/DM, but patients who died within 1 year after induction therapy were also included in this study. This study was approved by the Ethics Committee of Saitama Medical Center, Jichi Medical University (approved number 15–78) according to the Declaration of Helsinki.

Clinical evaluation

We retrospectively compiled the clinical data from the medical records, which included the age at disease onset, gender, disease duration, laboratory data, organ involvement (e.g. ILD, malignancy, cardiovascular disease and dysphagia), treatment history and disease activity after treatment. 'Severe muscle weakness' was defined as severe generalized muscle weakness due to myositis that necessitated assistance with activities of daily living. This definition was based on the myositis disease activity assessment tool provided by the International Myositis Assessment and Clinical Studies (IMACS) group [12]. The diagnosis of ILD was established by high-resolution chest CT according to the International Consensus Statement of Idiopathic Pulmonary Fibrosis of the American Thoracic Society [13]. RP-ILD was defined as the deterioration of radiological interstitial changes with symptoms of progressive dyspnoea and hypoxaemia within a few months from the onset of respiratory symptoms, and chronic-ILD was defined as the deterioration of radiological interstitial changes with symptoms of progressive dyspnoea for $>$3 months or no progression of radiological findings and/or respiratory symptoms for $>$3 months [6, 14].

Evaluation of serum MSAs

We comprehensively evaluated serum MSAs in 102 patients whose sera were available out of the 162 enrolled patients. Anti-ARS and anti-signal recognition particle (anti-SRP) antibodies were evaluated by RNA-immunoprecipitation assay using K562-cell extracts, as described previously [15]. Anti-MDA5 antibody was measured by ELISA [16]. Anti-Mi-2, anti-nuclear matrix protein 2 (anti-NXP2), anti-small ubiquitin-like modifier activating enzyme (anti-SAE) and anti-TIF1-γ were detected by immunoprecipitation-immunoblotting assay, as previously described [17, 18]. Anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase was not routinely evaluated in this study. The patients who lacked MSAs, including anti-ARS, anti-MDA5, anti-Mi-2, anti-NXP2, anti-SAE, anti-SRP and anti-TIF1-γ, were classified as MSA-negative in this study.

Definition of remission and sustained remission

Remission was defined as no evidence of disease activity, including cutaneous disease, muscle disease and pulmonary disease activities, in this study. The activities of the cutaneous disease, muscle disease and pulmonary disease were classified based on the Myositis Intention to Treat Activity Index (MITAX) scoring system [12]. The remission of the cutaneous disease was defined as no current activity of skin rash including the presence of Gottron's papules/sign, heliotrope rash and erythema, classified into the category D (currently inactive, but known to have been active in the past) according to the MITAX scoring system. The remission of the muscle disease was defined as a mean of $>$9 points on manual muscle testing by Kendall's 0–10-point scale for the neck and proximal muscles (neck flexor, deltoid, biceps, triceps, iliopsoas, quadriceps and hamstrings), $\geqslant$9 points for each one of these muscles [19], and no elevation of myogenic enzymes such as creatine kinase (CK) and aldolase, classified into the category D according to the MITAX scoring system. The remission of pulmonary disease was defined as no requirement of intensification of immunosuppressive treatment for ILD during the follow-up period, classified into the category C (stable disease) or D according to the MITAX scoring system.

Sustained remission was defined as a $>$6-month continuous period of the remission defined above while undergoing myositis therapy or no medication. This definition of sustained remission is almost identical to the definition of 'complete clinical response' by the IMACS group [20]. Sustained remission was retrospectively evaluated at either the most recent hospital visit or the date of death in medical chart.

Statistical analysis

All statistical analyses were performed using JMP 13 (SAS Institute Inc., Cary, NC, USA). A *P*-value <0.05 was regarded as significant. Fisher's exact test was used for binary data. Either the Mann–Whitney *U* test or the Kruskal–Wallis test was used for continuous data. Univariate and multivariate analyses were employed to find independent predictive factors for sustained remission. The multivariate analysis was performed by multivariate logistic regression analysis. Variables that were significant in the univariate analysis were selected as explanatory variables for the multivariate analysis. In this multivariate analysis, the presence of sustained remission was defined as an outcome variable. The odds ratio (OR) and CI were calculated per unit. The survival curve and cumulative remission rates were generated on Kaplan–Meier methods. The log-rank test was used to compare patient groups, and Bonferroni's correction was performed for multiple comparisons.

Results

Epidemiological and clinical characteristics of 162 patients with myositis at baseline

The baseline characteristics of all enrolled patients are shown in Table 1. The number of patients with PM, DM and CADM was 47 (29%), 85 (53%) and 30 (19%), respectively. Each clinical feature was noted as follows: dysphagia in 32 (20%), RP-ILD in 14 (9%), chronic-ILD in 74 (46%), malignancies in 40 (25%) and severe muscle weakness in 21 (13%). Regarding treatments, the median initial dose of prednisolone was 50 mg/day, which corresponded approximately to 1 mg/kg/day. Immunosuppressants were administered to 105 patients (65%) during the follow-up period. Immunosuppressants employed at induction therapy included MTX (18%), i.v. CYC (6%), calcineurin inhibitors (tacrolimus or ciclosporin A) (15%) and i.v. CYC combined with a calcineurin inhibitor (13%). IVIG was administered to 27 patients (17%). Three patients achieved remission by cancer treatment without immunosuppressive therapy. The median follow-up duration was 4 years.

Comparison of clinical features in each MSA group

We compared clinical features among MSAs in 102 patients whose sera were available (supplementary Table S1, available at *Rheumatology* online). The number of patients with anti-ARS, anti-MDA5, anti-NXP2, anti-SAE, anti-SRP and anti-TIF1-γ was 40 (39%), 15 (15%), 4 (4%), 2 (2%), 4 (4%) and 13 (13%), respectively. No patients with anti-Mi2 were observed in this study. There were 24 patients lacking MSAs (MSA-negative) (24%). The prevalence of dysphagia and malignancy was significantly higher in the patients with anti-TIF1-γ (54 and 77%, overall $P < 0.001$). RP-ILD was higher in patients with anti-MDA5 (27%), but it was not significant (overall $P = 0.31$). Chronic-ILD was significantly higher in patients with anti-ARS and anti-MDA5 (83 and 67%, overall $P < 0.001$). The levels of CK in patients with anti-MDA5 were significantly lower due to the high prevalence of CADM (overall $P = 0.001$). Most patients were prescribed immunosuppressants combined with prednisolone, but significantly fewer patients with anti-TIF1-γ received immunosuppressants (23%, overall $P < 0.001$). Significantly more patients with anti-SRP received IVIG (75%, overall $P = 0.041$).

Table 1 Baseline characteristics in enrolled patients

	n = 162
Age at disease onset, years	59 (44–67)
Female	115 (71)
PM, DM, CADM	47 (29), 85 (53), 30 (19)
Interval between onset and diagnosis, months	3 (1–7)
Clinical features	
Dysphagia	32 (20)
Heart involvement	3 (2)
RP-ILD	14 (9)
Chronic-ILD	74 (46)
Malignancy	40 (25)
Severe muscle weakness	21 (13)
Initial laboratory data at disease onset	
CK, IU/L	551 (133–2383)
CRP, mg/dL	0.7 (0.2–2.4)
Treatment	
Initial dose of PSL, mg/day	50 (40–60)
PSL + immunosuppressants	105 (65)
Immunosuppressants at induction therapy	
MTX	29 (18)
I.v. CYC	10 (6)
Calcineurin inhibitors	25 (15)
I.v. CYC + calcineurin inhibitors	21 (13)
IVIG	27 (17)
Follow-up period, years	4 (1–6)

Data are expressed as *n* (%) or median (interquartile range). CDAM: clinically amyopathic dermatomyositis; Chronic-ILD: chronic interstitial lung disease; CK: creatinine kinase; PSL: prednisolone; RP-ILD: rapidly progressive interstitial lung disease.

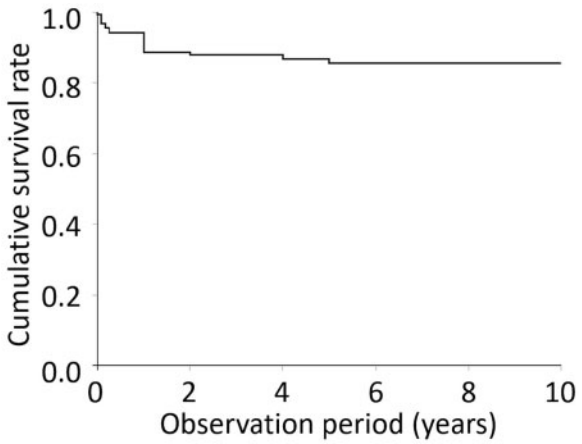
Prognosis and cause of death in all enrolled patients

The survival rates were 89, 86 and 86% at 1, 5 and 10 years, respectively (Fig. 1). Twenty-six patients (16%) died during the follow-up period. The causes of deaths were as follows: malignancy in 10 (38%), infectious pneumonia in 6 (23%), RP-ILD in 5 (19%), subarachnoid haemorrhage in 2 (8%), heart failure in 1 (4%), suicide in 1 (4%) and unknown cause of death in 1 patient (4%).

Achievement rates of sustained remission stratified by MSAs

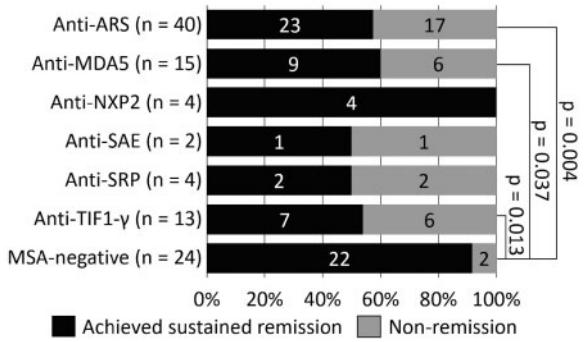
Among all 162 patients, 94 patients achieved sustained remission (58%). The number of patients who achieved sustained remission stratified by each MSA is shown in Fig. 2 (overall $P = 0.022$). Pairwise comparisons demonstrated that the achievement rate of sustained remission

Fig. 1 Cumulative survival rate of all enrolled patients



The survival curve was generated based on Kaplan–Meier methods.

Fig. 2 Achievement rates of sustained remission stratified by MSAs



Non-remission means the patients who did not achieve sustained remission. The overall *P*-value was 0.022. Pairwise comparisons: $P = 0.004$ between MSA-negative and anti-ARS, $P = 0.037$ between MSA-negative and anti-MDA5, and $P = 0.013$ between MSA-negative and anti-TIF1-γ. Anti-ARS: anti-aminoacyl-transfer ribonucleic acid synthetase; anti-MDA5: anti-melanoma differentiation-associated gene 5; anti-NXP2: anti-nuclear matrix protein 2; anti-SAE: anti-small ubiquitin-like modifier activating enzyme; anti-SRP: anti-signal recognition particle; anti-TIF1-γ: anti-transcriptional intermediary factor 1-γ; MSA: myositis-specific autoantibody.

for MSA-negative patients was significantly higher than that for patients with anti-ARS ($P = 0.004$), anti-MDA5 ($P = 0.037$) and anti-TIF1-γ ($P = 0.013$). Sustained remission rates stratified by each MSA after exclusion of patients who died during the follow-up period are shown in supplementary Fig. S1, available at *Rheumatology* online. The results were almost identical to those without excluding the patients who died.

Predictive factors associated with sustained remission

Predictive factors associated with sustained remission in the patients with PM/DM were analysed. The results of univariate logistic regression analysis are presented in Table 2. Female gender, age at disease onset, dysphagia, RP-ILD, chronic-ILD, malignancy, severe muscle weakness, MSA-negative, combination with immunosuppressants, combination with IVIG and the follow-up period were significantly associated with sustained remission in univariate analyses. Multivariate logistic regression analysis demonstrated that the absence of severe muscle weakness (OR 43.6; 95% CI 4.97, 1126; $P < 0.001$) and MSA-negative (OR 5.84; 95% CI 1.51, 31.4; $P = 0.009$) were both independent predictive factors associated with sustained remission.

Cumulative remission rates based on stratification by predictive factors for sustained remission

We also analysed cumulative remission rates based on Kaplan–Meier methods to compare the periods of sustained remission stratified by the two predictive factors regarding the achievement of sustained remission (factor 1: the absence of severe muscle weakness; and factor 2: MSA-negative status) revealed by multivariate logistic regression analysis (Fig. 3). The sustained remission periods were calculated from the achievement of remission after the first induction therapy to the date of the most recent hospital visit, recurrence or death. Forty-nine patients who had never reached remission during the follow-up period were excluded. The cumulative remission rates for all patients in this analysis ($n = 113$) are shown in Fig. 3A, demonstrating the 10-year remission rate of 54%. The cumulative remission rates were significantly higher for patients with factor 1 ($P < 0.001$) (Fig. 3B) and patients with factor 2 ($P < 0.001$) (Fig. 3C) than the others. The cumulative remission rates were notably higher in patients with both factor 1 and factor 2, as shown in Fig. 3D (overall $P < 0.001$).

The cumulative remission rates stratified by MSAs are shown in Fig. 4. This investigation also revealed that MSA-negative patients had significantly higher cumulative remission rates (overall $P < 0.001$; pairwise comparisons: $P < 0.001$ between MSA-negative and anti-SRP, and $P = 0.005$ between MSA-negative and anti-ARS).

Discussion

This study clarified the sustained remission rate, and demonstrated the absence of severe muscle weakness requiring assistance at diagnosis and MSA-negative status to be independent predictive factors for sustained remission in adult patients with PM/DM using our large database of enrolled patients from a single cohort. To our knowledge, this is the first study to clarify the predictive factors for sustained remission in adult PM/DM in daily practice.

Previous reports regarding remission rates in PM/DM are limited, and the definition of remission has not been

Table 2 Predictive factors associated with sustained remission by univariate analysis and multivariate analysis

	Univariate analyses			Multivariate analyses*		
	Odds ratio	95% CI	*P*-value	Odds ratio	95% CI	*P*-value
Female	2.15	1.08, 4.32	0.028	0.32	0.07, 1.25	0.10
Age at disease onset	0.98	0.96, 1.00	0.041	1.01	0.97, 1.05	0.62
Interval between onset and diagnosis	0.85	0.64, 1.03	0.11			
PM	0.86	0.43, 1.71	0.66			
DM	1.31	0.70, 2.46	0.39			
CADM	0.79	0.36, 1.77	0.57			
Dysphagia	0.30	0.13, 0.66	0.003	2.95	0.32, 71.1	0.37
Heart involvement	0.35	0.02, 3.78	0.38			
RP-ILD	0.26	0.07, 0.81	0.020	0.21	0.02, 1.58	0.13
Chronic-ILD	2.08	1.10, 4.00	0.023	2.02	0.56, 7.43	0.28
Malignancy	0.43	0.21, 0.89	0.023	2.15	0.42, 14.5	0.37
Absence of severe muscle weakness	10.9	3.49, 48.2	<0.001	43.6	4.97, 1126	<0.001
CK levels	1.00	1.00, 1.00	0.22			
Anti-ARS	0.54	0.23, 1.24	0.15			
Anti-MDA5	0.74	0.24, 2.38	0.60			
Anti-NXP2	$5.9*10^6$	0.86, 0.00	0.065			
Anti-SAE	0.51	0.02, 13.1	0.64			
Anti-SRP	0.50	0.06, 4.31	0.50			
Anti-TIF1-γ	0.55	0.17, 1.86	0.33			
MSA-negative	5.10	1.60, 22.8	0.005	5.84	1.51, 31.4	0.009
Initial dose of PSL	1.00	0.99, 1.00	0.38			
Combination with immunosuppressants	1.96	1.02, 3.79	0.043	1.51	0.34, 6.26	0.57
Combination with IVIG	0.29	0.12, 0.69	0.005	0.51	0.12, 2.09	0.34
Follow-up period	1.14	1.06, 1.25	<0.001	2.63	0.93, 1.15	0.59

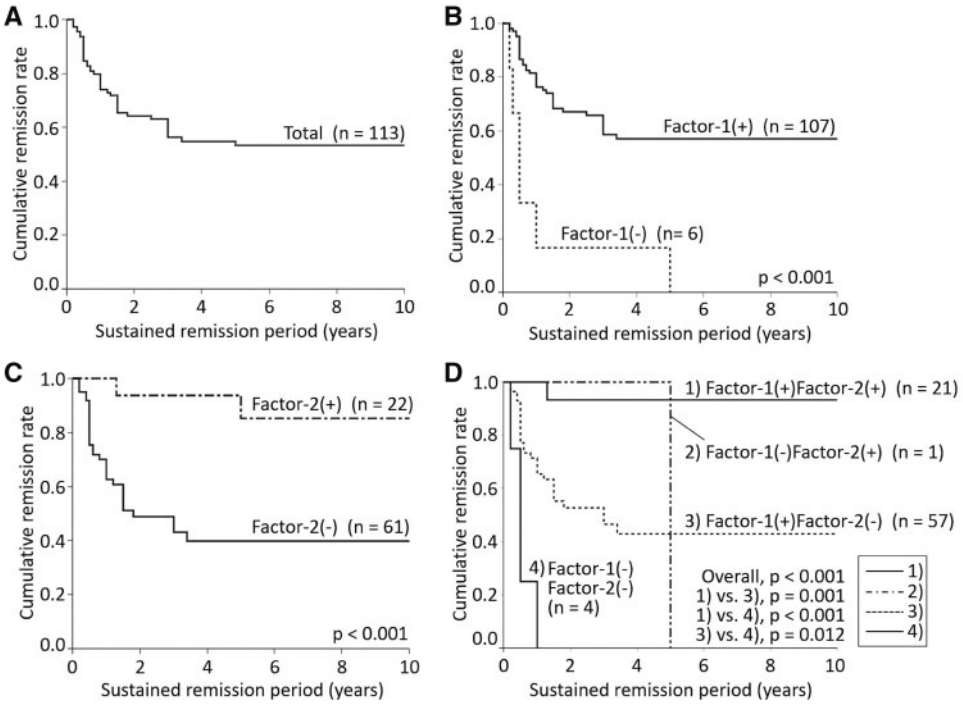
*Variables that were significant (*P*-value <0.05) in the univariate analysis were selected as explanatory variables for the multivariate analysis. In this multivariate analysis, the presence of sustained remission was defined as an outcome variable. anti-ARS: anti-aminoacyl-transfer ribonucleic acid synthetase; anti-MDA5: anti-melanoma differentiation-associated gene 5; anti-NXP2: anti-nuclear matrix protein 2; anti-SAE: anti-small ubiquitin-like modifier activating enzyme; anti-SRP: anti-signal recognition particle; anti-TIF1-γ: anti-transcriptional intermediary factor 1-γ; CDAM: clinically amyopathic dermatomyositis; Chronic-ILD: chronic interstitial lung disease; CK: creatinine kinase; MSA: myositis-specific autoantibody; PSL: prednisolone; RP-ILD: rapidly progressive interstitial lung disease.

consistent throughout previous studies. The remission rates depend on definitions of remission for PM/DM. Bronner *et al.* defined remission as no detectable clinical or biochemical disease activity and absence of drugs, and reported a remission rate of 20% [21]. Other retrospective cohort studies reported remission rates of 15.2% [22] and 57.3% [23], but the definitions of remission were not described in detail. In this study, we clarified the sustained remission rate at ~60% using a clear definition of sustained remission based on the MITAX scoring system and the international consensus guidelines for trials of myositis therapies defined by the IMACS group [12, 20], although our definition was not consistently approved and validated across the world. In addition, PM/DM patients with stable disease of ILD, identical to the category C in the MITAX scoring system, were classified into the remission group as well as PM/DM patients with currently inactive disease in this study. In the future, the definition of remission for PM/DM that is applicable in daily practice needs to be determined.

In this study, we identified the absence of severe muscle weakness requiring assistance at diagnosis as one of the independent predictive factors for sustained remission. Severe muscle weakness requiring assistance can be objectively evaluated, and quickly distinguishes between patients with and without a favourable outcome in daily practice. A previous cohort study reported that a significant degree of muscle weakness on presentation does not preclude remission in JDM [9]. However, muscle weakness is often severe in adult PM/DM patients with malignancy and/or dysphagia [24–28]. In our study, 11 patients (52%) with severe muscle weakness developed malignancy and 15 patients (71%) presented dysphagia. The presence of malignancy and/or dysphagia is a poor prognostic factor for PM/DM [4]; thus, these complications may lead to the failure of sustained remission in patients with severe muscle weakness.

MSA-negative status was the other independent predictive factor for sustained remission in PM/DM patients. In addition, the remission rate remained high during the follow-up period in the MSA-negative group (Figs 3C and 4), suggesting that long-term sustained remission is possible after induction therapy in this group. Little is known about the clinical features and outcomes of MSA-negative

FIG. 3 Cumulative remission rates stratified by predictive factors associated with achievement of sustained remission

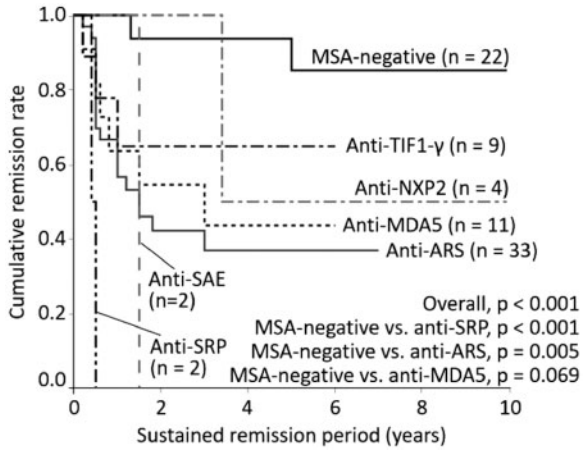


Cumulative remission rates were generated based on Kaplan–Meier method. Forty-nine patients who had never reached remission during the follow-up period were excluded. Two predictive factors associated with achievement of sustained remission were as follows: factor 1: the absence of severe muscle weakness requiring assistance; and factor 2: myositis-specific autoantibody-negative status. Cumulative remission rates for all patients (**A**), a comparison of the cumulative remission rates between patients with and without factor 1 (**B**), a comparison of the cumulative remission rates between patients with and without factor 2 among 83 patients in whom MSAs were evaluated (**C**), and comparisons of the cumulative remission rates between four groups: group 1, patients with both factor 1 and factor 2; group 2, patients with factor 2 alone; group 3, patients with factor 1 alone; and group 4, patients having neither factor 1 nor factor 2 (**D**). MSA: myositis-specific autoantibody.

patients. The clinical features of MSA-negative patients were reported to be mild-to-moderate muscle involvement with classical skin rash [29, 30]. In our study, muscle involvement in MSA-negative patients was mild-to-moderate because of moderate CK levels and lower prevalence of severe muscle weakness (supplementary Table 1, available at *Rheumatology* online). Skin rash was observed in 63% of MSA-negative patients, but it was not severe. The lower severity of skin and muscle involvement might have contributed to the better outcome for MSA-negative patients. The incidence and severity of lung involvement in MSA-negative patients are also unknown [29, 30]. Our study revealed that 58% of MSA-negative patients had ILD, of which the major subtype was chronic-ILD, and the clinical outcome was significantly better than that in a previous cohort study [31]. In our study, 86% of MSA-negative patients with ILD received immunosuppressants, hence the higher rate of concomitant immunosuppressant use also might be attributed to the better outcome. Regarding malignancy, there were four malignancies (17%) among the MSA-negative patients. Malignancy develops less frequently in these patients than those with anti-TIF1-γ [32]. Further studies are required to clarify the clinical characteristics and outcomes of MSA-negative patients.

This study had several limitations. First, this study was a retrospective analysis conducted at a single institute. There may have been a patient-selection bias because of the retrospective design; however, the distribution of idiopathic inflammatory myopathy type, sex ratio, age, and incidence of malignancies and organ involvement in our study were almost identical to those in previous studies. Second, anti-Mi-2, anti-NXP2, anti-SAE and anti-SRP were detected in no or few patients in this study, which may have affected the results. Third, we did not routinely examine for anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase antibody, although one PM patient with statin-induced myositis suspected to have anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase antibody was excluded from the study. Fourth, rituximab is not approved by the Japanese government for myositis patients on national health insurance, as of June 2019. Therefore, there was no patient who received rituximab

Fig. 4 Cumulative remission rates stratified by MSAs in patients with PM/DM



Cumulative remission rates were generated based on Kaplan–Meier method for patients in whom MSAs were evaluated. Nineteen patients who had never reached remission during the follow-up period were excluded. Anti-ARS: anti-aminoacyl-transfer ribonucleic acid synthetase; anti-MDA5: anti-melanoma differentiation-associated gene 5; anti-NXP2: anti-nuclear matrix protein 2; anti-SAE: anti-small ubiquitin-like modifier activating enzyme; anti-SRP: anti-signal recognition particle; anti-TIF1-γ: anti-transcriptional intermediary factor 1-γ; MSA: myositis-specific autoantibody.

in this study. This differs from western countries. The interpretation of our result should be cautious. Fifth, data were lacking on disease activity evaluated by the physician global activity and patient global activity, and extramuscular disease activity, except for pulmonary disease provided by the IMACS group [12, 33].

In conclusion, the absence of severe muscle weakness requiring assistance at diagnosis and MSA-negative status are independent predictive factors for sustained remission in adult PM/DM patients. These predictive factors should be validated in a multicentre prospective cohort in the future to improve treatment strategies and outcomes for patients with PM/DM.

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Supplementary data

Supplementary data are available at *Rheumatology* online.

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